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Case report

Treatment of superior oblique myokymia with oxcarbazepine

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ABSTRACT

A 33-year-old male patient with ipsilateral intermittent oscillopsia and blurry vision for 18 months was found to have intorsional nystagmus of the right eye, especially when gazing at the nasal lower field. The oscillations improved with exercise and were aggravated by rest. An abnormal head posture, with a tilt towards the left, and mild right superior oblique muscle paresis were noted by the prism cover test and motility examination. Neurovascular compression was not confirmed by 0.9-mm thick, high-resolution magnetic resonance imaging with fast imaging employing steady state acquisition sequence. After treatment with oxcarbazepine (600 mg/day), the oscillations resolved and there have been no complications during the 1-year follow-up period. Superior oblique myokymia is a rare form of nystagmus that may cause oscillopsia and blurry vision. Oxcarbazepine, a structural derivative of carbamazepine, can be used to successfully eliminate oscillopsia without serious adverse reaction.

1. Introduction

Superior oblique myokymia (SOM) is an uncommon disorder; it typically presents with intermittent ipsilateral oscillopsia, first described by Duane[1] in 1906. The pathogenesis of this disease entity is not fully understood. However, neurovascular compression of the trochlear nerve may play an important role.[2] Medical treatment,[3] extraocular surgery of involved muscles,[4] and neurovascular decompression of the trochlear nerve[5] have been reported as treatments for SOM. We report a case of SOM without definite magnetic resonance imaging evidence of vascular compression; the symptoms were successfully controlled using oxcarbazepine.

2. Case report

A 33-year-old male patient presented to our clinic with unilateral intermittent oscillopsia and double vision for 18 months. The visual symptoms were reported to partially improve with exercise and extreme fatigue, but were aggravated at rest. The position of the head was tilted to the left, which helped alleviate visual symptoms. The patient had a corrected visual acuity of 20/20 in both eyes. Motility examination showed mild updrift of the right eye on levoversion. The prism cover test revealed small angle right hypertropia of approximately five prism diopters. Intorsional twitching of the right eye was observed under slit lamp examination. Intorsional nystagmus was more noticeable with the gaze to the nasal lower field of the right eye (Fig. 1). In 0.9-mm thickness cut, T2-weighted magnetic resonance imaging with fast imaging employing steady state acquisition (FIESTA) sequence at his subarachnoid area, the proximal portion of the right trochlear nerve could not be identified as compared to the visible nerve on the left side (Fig. 2A). In 1-mm thickness cut, three-dimensional (3D) time of flight magnetic resonance angiography, the path of the right medial superior cerebellar artery (SCA) was similar to the left trochlear nerve identified in magnetic resonance imaging (Fig. 2B). Although evidence of neurovascular compression of the right trochlear nerve was not found, we started medical treatment under the clinical diagnosis of SOM.

Oxcarbazepine, 600 mg/day, was prescribed and the symptoms resolved. The patient remained symptom-free with no torsional nystagmus observed during the follow-up period of 1 year.

3. Discussion

SOM is an uncommon disorder characterized by brief episodes of unilateral oscillopsia and/or diplopia. The term was introduced by Hoyt and Keane[6] in 1970. The associated symptoms are thought to be caused by abnormal electrical activity of the trochlear nerve and twitching of the superior oblique muscle, leading to intorsional

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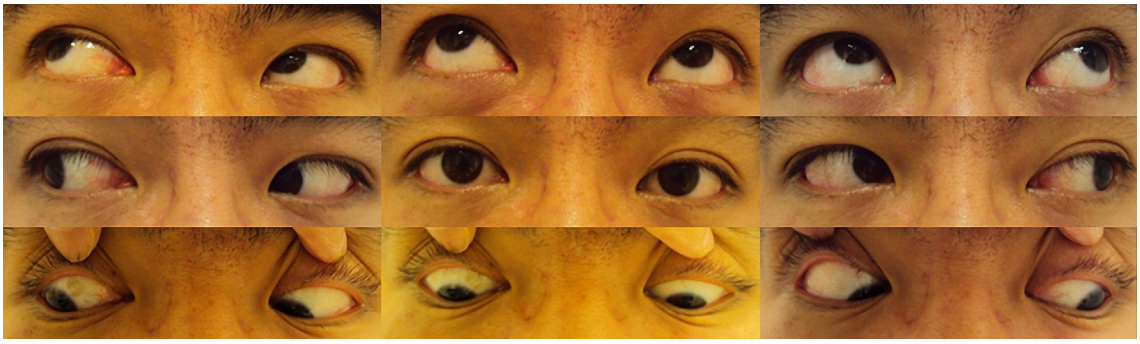

Fig. 1. Motility examination showed right hypertropia at primary position and mild over-elevation in adduction in the right eye of the patient. The patient had torsional oscillopsia especially in the field of gaze of the right superior oblique muscle.

nystagmus of the globe. In most cases of SOM, the etiology of the abnormal activity of the trochlear nerve remains unconfirmed. One of the postulated hypotheses to explain the associated pathology is vascular compression of the cistern segment of the trochlear nerve, leading to damage of the area between the central and peripheral myelin, which is particularly vulnerable to pulsatile pressure, causing abnormal muscle twitching as in hemi-facial spasm.[7] The long intracranial course (60 mm) and narrow diameter (0.2–0.6 mm)[8,9] of the trochlear nerve makes it fragile and easily affected by surrounding structures.

Imaging the cistern segment of the trochlear nerve has been a challenge because of its small diameter. The proximity of the trochlear nerve to nearby vessels taking a similar course and with similar caliber has also increased the difficulty in determining the specific neurovascular relationships. Casselman et al[10] postulated 3D constructive interference in steady state (CISS) magnetic resonance imaging with a section thickness of 0.7 mm to image the cisternal portion of the trochlear nerve in 1994. Yousry et al[11] successfully depicted the anatomy of the trochlear nerve in 30 volunteers and a patient with SOM using 0.66-mm thickness magnetic resonance imaging with 3D CISS sequences. The detection rate was highest at transverse plane, reaching 95% among the 60 nerves imaged. With utilization of 3D balanced turbo field echo sequence at the pontomesencephalic junction, section thickness of 0.25 mm, Yang et al[12] described the detailed anatomy of the cisternal segment of the trochlear nerve in 97 superior oblique palsy patients. One of the reasons that we failed to identify neurovascular contact might be the effective section thickness used. We scanned the patient with cut thickness of 0.9 mm, which was more than 0.7 mm suggested by a previous study.[10] However, neurovascular contact is not a marker for diagnosing SOM because it is not a rare anatomic phenomenon. Abducens nerve was found to be in contact with the anterior inferior cerebellar artery in 76.6% of cases in a magnetic resonance imaging study.[13] Yousry et al[11] found that medial SCA branches were in direct contact with 51% of the trochlear nerve identified. At present, diagnosis of SOM still mainly depends on clinical symptoms and signs.

Various medications have been used to treat SOM. Carbamazepine, a neural membrane stabilizing agent, is the most frequently administered medication. Its use has shown promising treatment results.[3,14,15], Benefits from phenytoin, baclofen, propranolol, and clonazepam have also been reported.[16,17] Williams et al[3] reported that 15 out of 18 (83%) patients with SOM benefited from carbamazepine, 400 mg/day, titrated to 800 mg/day as tolerated. Six patients (30%) stopped treatment because of adverse effects such as rash, elevated liver enzyme, mild nausea, and/or lethargy. Surgical treatments are alternatives when medical treatment is unsuccessful. Surgery of the extraocular muscles with the combination of superior oblique tenectomy and inferior oblique myectomy has been reported to be effective in eliminating oscillopsia associated with SOM;

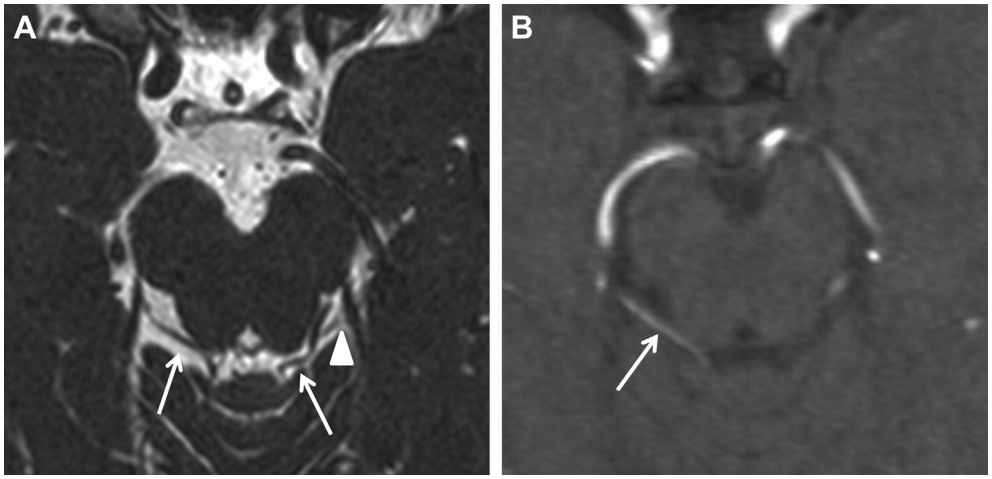

Fig. 2. (A) Magnetic resonance imaging with fast imaging employing steady state acquisition (FIESTA) sequence, constructive interference in steady state (CISS) T2 image, in the subarachnoid area of the patient with SOM. The cut thickness was 0.9 mm (TE 5.47). The left trochlear nerve (arrowhead) and medial superior cerebellar artery (SCA) branches (arrows) can be seen. The right medial SCA is overlapped at the root exit zone and right trochlear nerve cannot be clearly distinguished from the artery on the right side. (B) One-mm cut, 3D transverse time of flight magnetic resonance angiogram (TE 7.2) confirmed that the structure at the root entry zone is an artery (arrow).

however, diplopia in downward gaze was noted in approximately 36% of patients.[4] Although treatment success has been reported,[5] the risks of intracranial microvascular decompression surgery probably outweigh the benefits in most cases.

Oxcarbazepine is a derivative of carbamazepine. It is a pro-drug and is activated to eslicarbazepine in the liver.[18] Like carbamazepine, it works as an inhibitor of the voltage-dependent sodium channel and decreases the excitability of the cell membrane. The structural differences from carbamazepine reduce the metabolic effects on the liver and decreases hematological side effects occasionally associated with carbamazepine. The patient reported here responded well to oxcarbazepine and was symptom-free without adverse effects at 1 year follow-up.

In conclusion, a rare case of SOM is reported without definite imaging evidence of neurovascular contact. With diagnosis based upon clinical presentations, visual symptoms can be successfully treated with oxcarbazepine.

References

1. Duane A. Unilateral rotary nystagmus. *Trans Am Ophthalmol Soc*. 1906;11:63–67.
2. Hashimoto M, Ohtsuka K, Hoyt WF. Vascular compression as a cause of superior oblique myokymia disclosed by thin-slice magnetic resonance imaging. *Am J Ophthalmol*. 2001;131:676–677.
3. Williams PE, Purvin VA, Kawasaki A. Superior oblique myokymia: efficacy of medical treatment. *J AAPOS*. 2007;11:254–257.
4. Ferreira T, Verbist B, van Buchem M, van Osch T, Webb A. Imaging the ocular motor nerves. *Eur J Radiol*. 2010;74:314–322.
5. Samii M, Rosahl SK, Carvalho GA, Krzizok T. Microvascular decompression for superior oblique myokymia: first experience. Case report. *J Neurosurg*. 1998;89:1020–1024.
6. Hoyt WF, Keane JR. Superior oblique myokymia. Report and discussion on five cases of benign intermittent uniocular microtremor. *Arch Ophthalmol*. 1970;84:461–467.
7. Hashimoto M, Ohtsuka K, Suzuki Y, Minamida Y, Houkin K. Superior oblique myokymia caused by vascular compression. *J Neuroophthalmol*. 2004;24:237–239.
8. Villain M, Segnarbieux F, Bonnel F, Aubry I, Arnaud B. The trochlear nerve: anatomy by microdissection. *Surg Radiol Anat*. 1993;15:169–173.
9. Ettl A, Salomonowitz E. Visualization of the oculomotor cranial nerves by magnetic resonance imaging. *Strabismus*. 2004;12:85–96.
10. Casselman JW, Kuhweide R, Dehaene I, Ampe W, Devlies F. Magnetic resonance examination of the inner ear and cerebellopontine angle in patients with vertigo and/or abnormal findings at vestibular testing. *Acta Otolaryngol Suppl*. 1994;513:15–27.
11. Yousry I, Moriggl B, Dieterich M, Naidich TP, Schmid UD, Yousry TA. MR anatomy of the proximal cisternal segment of the trochlear nerve: neurovascular relationships and landmarks. *Radiology*. 2002;223:31–38.
12. Yang HK, Kim JH, Hwang JM. Congenital superior oblique palsy and trochlear nerve absence: a clinical and radiological study. *Ophthalmology*. 2012;119:170–177.
13. Yousry I, Camelio S, Wiesmann M, et al. Detailed magnetic resonance imaging anatomy of the cisternal segment of the abducent nerve: Dorello's canal and neurovascular relationships and landmarks. *J Neurosurg*. 1999;91:276–283.
14. Susac JO, Smith JL, Schatz NJ. Superior oblique myokymia. *Arch Neurol*. 1973;29:432–434.
15. Rosenberg ML, Glaser JS. Superior oblique myokymia. *Ann Neurol*. 1983;13:667–669.
16. Staudenmaier C. Superior oblique myokymia: when treatment is necessary. *Can J Ophthalmol*. 1986;21:236–241.
17. Tyler TD, Ruiz RS. Propranolol in the treatment of superior oblique myokymia. *Arch Ophthalmol*. 1990;108:175–176.
18. Dulsat C, Mealy N, Castaner R, Bolos J. Eslicarbazepine acetate. *Drugs Future*. 2009;34:189.